

# Modern Concepts of Cardiovascular Disease

Published monthly by the AMERICAN HEART ASSOCIATION

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COLUMBIAN UNIVERSITY

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Vol. X

February, 1941

No. 2

## ANEURYSM OF THE TRUNK AND MAIN BRANCHES OF THE PULMONARY ARTERY: ANALYSIS OF 152 CASES

### INTRODUCTION

There appears to be some confusion as to what material should be included and what excluded in a discussion of aneurysm of the pulmonary artery. If one holds strictly to the definition of aneurysm as a dilatation of an artery as the result of, or in association with, actual damage to one or more of its layers, at least 116 instances of aneurysm of the trunk and/or main branches of the pulmonary artery confirmed at autopsy can be collected from the literature, and thirty-five additional instances can be accepted on purely clinical grounds. Excluded from this grouping are instances of diffuse dilatation of the pulmonary artery and its branches, all dissecting aneurysms, save those which arise on the basis of previous aneurysmal dilatation, and all aneurysms of the smaller branches of the pulmonary artery.

### PATHOGENESIS

The differences in the development and course of aneurysms of the pulmonary artery and those arising in the major circulation may be readily appreciated if it is remembered that:

1. Congenital anomalies are common in connection with the former.
2. The pulmonary artery is thinner than other vessels of equal calibre, being particularly deficient in elastic and muscular fibers in the media.
3. The elastic and muscular component of the media is particularly lacking along the anterior and lateral walls of the truncus.
4. The pulmonary artery is easily distensible, presumably because of the low tonus of all vessels in the pulmonary circuit.
5. The pulse pressure in the lesser circulation exceeds the mean pulmonary arterial pressure.

### LOCATION OF THE ANEURYSM

Of the recorded cases confirmed by autopsy, 80 per cent (93 cases) involve the trunk of the artery, of which 14 involve one or both branches as well. The right or left main branches or both are affected in 23 instances. Of all these aneurysms, slightly more than half (57.8 per cent) are sacular.

### AGE AND SEX INCIDENCE

The disease is equally distributed between the sexes (57 males and 56 females) (Table I). The average age is 37.7 years (Table I) being approximately the same for the members of each sex. Two distinct age peaks may be seen; the first from birth to the 29th year, and the second from the 40th to the 59th years. The former represents those instances of the disease occurring on the basis of congenital anomalies, the latter those individuals with predominantly arteriosclerotic or, more rarely, syphilitic lesions.

### ETIOLOGY

The etiologic factors are tabulated in Table I. Congenital anomalies, present in at least 66 per cent of the cases, were looked upon as predisposing causes in 43.2 per cent of the patients. Among the precipitating factors in this group were pulmonary hypertension, atheromatosis, and infection. Twenty-three per cent of 104 cases showed arteriosclerosis of the pulmonary artery of sufficient degree to be of etiologic significance. Syphilis appears to have been present and active in 31.7 per cent. Other in-

fections (17.2 per cent) have been observed to initiate the changes giving rise to aneurysms of the pulmonary artery, particularly where congenital anomaly exists.

### CLINICAL FEATURES

Patients with aneurysm of the pulmonary artery may be divided clinically into two main groups, viz., those in whom the trunk, alone or in combination, is involved, and those in whom one or both branches are affected. The former type is recognizable on the basis of signs and symptoms, whereas the latter gives rise to symptoms alone. Syphilis seems to be of slightly more frequent occurrence in the latter than in the former, whereas congenital lesions are equally distributed.

Many complicating and associated pathologic conditions lend variety to the clinical picture (Table II), by no means the least of which are interauricular defects and the changes in the pulmonary valve and artery itself.

Antemortem diagnosis is difficult in both clinical types, but can be established with a fair degree of accuracy in the first group if the following are simultaneously present: stasis in the lesser circulation, characterized by dyspnea, cyanosis, oppression of the chest, and bloody sputum; prominence of the left side of the chest in the region of the second and third costal cartilages; a pulsation, a thrill, and a loud superficial, sawing or rubbing systolic murmur, best heard at the second left intercostal space; a weak cardiac thrust with dullness not going beyond the mid-clavicular line; hypertrophy and dilatation of the right side of the heart; electrocardiographic evidence of right axis deviation; roentgenographic demonstration of lengthening and lateral projection of the pulmonary conus shadow in the anterior view, with a see-saw movement of the left border; indentation of the oesophagus below the normal aortic impression in the right anterior oblique position; and encroachment upon the ventral and caudal aspects of the aortic window in the left anterior oblique visualization.

The subjective symptoms are similar in the two groups. Cough occurs in 80 per cent of the cases, but is rarely an early manifestation. It depends upon pressure on the recurrent laryngeal nerve, in which event it is non-productive; upon bronchial irritation with at first a dry, and later a productive phase; or upon pulmonary thromboses when it is blood streaked or frankly hemorrhagic.

Dyspnea appears in 75 to 80 per cent of all patients, and together with cyanosis, is seen early in the congenital type, whereas precordial, shoulder or arm pain, and cough antedate them in the acquired form. Peripheral edema is rare, but pulmonary stasis is constant, except in lesions, usually syphilitic, affecting but a single large branch of the pulmonary vessel.

Physical findings of value are present only when the truncus is involved. In 44 of 48 patients, the left side of the chest, particularly in the region of the second and third costal cartilages was more prominent than the right. Pulsation (31 instances) with systolic (28 cases) or diastolic (3 cases) fremitus was noted in the second left costal interspace in 31 of 48 patients. Enlargement of the cardiac contour was clinically demonstrable in the left second

interspace in 25 of 48 patients, and to the right of the sternum in 12. A systolic murmur was present in 85 per cent in the second and third left intercostal spaces, and diastolic bruit, usually indicating complicating infection, in 12 per cent.

#### DIFFERENTIAL DIAGNOSIS

A number of conditions may simulate the clinical picture of aneurysm of the pulmonary artery:

1. Aortic aneurysm. Roentgenologic examination will usually differentiate the shadow cast by a lesion of the descending aorta from one resulting from aneurysm of the pulmonary artery. In the difficult case resort may be had to the Robb-Steinberg technique.

2. Patent ductus arteriosus. Differentiation on clinical grounds is impossible when aneurysm or thrombosis of the duct is present. Either one of these conditions may complicate the course of pulmonary artery disease.

3. Idiopathic dilatation of the pulmonary artery. The "hilar dance" is particularly prominent, and the denseness of the large arteries with the "comma-

like" appearance of the right branch of the pulmonary artery usually leaves little room for confusion.

4. Interauricular septal defects. The absence of murmurs, the right axis deviation, the late cyanosis, and "paradoxical embolism" serve to distinguish this lesion.

5. Other congenital heart lesions. As a rule the prominence of the chest, the character of the pulsation, the peculiar thrill and murmur of aneurysm of the pulmonary artery serve to differentiate it from other anomalies of the heart with the exception of patent ductus arteriosus.

#### SUMMARY

The clinical picture of aneurysm of the pulmonary artery as reconstructed from a study of 151 recorded instances of that disease, has been briefly summarized, with special emphasis upon etiologic, age, sex, roentgenographic, and differential diagnostic factors.

Linn J. Boyd, M.D.

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**TABLE I**  
**RELATION OF AGE, SEX, AND ETIOLOGIC FACTORS IN 113 CASES OF ANEURYSM OF THE PULMONARY ARTERY**

No. Cases	Etiology	Percentage total	Age			Sex					
			Average	Max.*	Min.*	Male	Percent	Average age	Female	Percent	Average age
113		100	37.7	F 82	F 4	57	50.4	37.2	56	49.6	38.7
3	Trauma	2.8	32.3	—	—	—	—	—	—	—	—
45	Congenital defect	43.2	31.8	F 82	F 4	20	44.5	31.0	25	55.5	32.0
24	Arterio-sclerosis	23.0	47.5	F 79	M 22	13	54.1	43.0	11	45.9	52.7
33	Syphilis	31.7	45.7	F 73	F 19	15	50.0	41.7	15	50.0	46.9
7	Rheumatism	6.7	35.0	F 54	M 22	2	28.6	23.5	5	71.4	39.8
6	Septic endocarditis or endarteritis	5.7	26.8	F 35 M 35	F 13	2	40.0	32.5	3	60.0	23.0
5	Infection—type not specified	4.8	21.2	F 37	F 13	1	20.0	15.0	4	80.0	22.7

\* F—Female; M—Male, in these columns

**TABLE II**  
**ASSOCIATED PATHOLOGIC CONDITIONS OR COMPLICATIONS IN 104 CASES OF ANEURYSM OF THE PULMONARY ARTERY**

Location and Condition		No. Cases	% Total	Location and Condition		No. Cases	% Total
<b>Aorta</b>		40	38.4	<b>Pericardium</b>		9	8.6
Hypoplasia	6	5.7		Rheumatic Pericarditis	3	2.9	
Congenital Stenosis	7	6.7		Adhesive Pericarditis	2	1.9	
Coarctation	1	.96		Not specified	4	3.8	
Atheroma	5	4.8		<b>Pulmonary Artery</b>		65	62.5
Syphilis	21	21.9		Congenital Stenosis	4	3.8	
with aneurysm	5	4.8		Hypoplasia	2	1.9	
with dilatation	1	.96		Widespread Sclerosis (non-specific)	24	23.8	
<b>Mitral Valve</b>		12	11.5	Syphilis	33	31.7	
Congenital Stenosis	2	1.9		Septic Endarteritis	2	1.9	
Endocarditis with Stenosis	9	8.6		<b>Congenital Defects (Not Previously Mentioned)</b>		39	37.5
Rheumatic	6	5.7		Inter-auricular	11	10.5	
Luetic (?)	1	.96		Inter-ventricular	4	3.8	
Not Specified	2	1.9		Patent ductus botalli with vegetatives	24	23.1	
Insufficiency	1	.96		with aneurysm	3	2.9	
<b>Tricuspid Valve Stenosis</b>		1	.96		1	.96	
<b>Pulmonary Valve</b>		24	23.8	<b>Infections (Not Otherwise Mentioned)</b>		18	17.3
Bi-cuspid	1	.96		Pulmonary Tuberculosis	9	8.6	
Quadri-cuspid	1	.96		Pulmonary Syphilis	1	.96	
Congenital Stenosis	12	11.5		Endocarditis	4	3.8	
Insufficiency	2	1.9		Cardiac Lues	2	1.9	
Sclerosis with Stenosis	1	.96		Rheumatic Pancarditis	1	.96	
Endocarditis	6	5.7		Actinomycosis Lungs and Pleura	1	.96	
Luetic	3	2.9					
Aneurysm	1	.96					

